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Unilateral pulmonary agenesis and esophageal atresia with a tracheoesphageal fistula-23 year followup

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ABSTRACT

Pulmonary agenesis, esophageal atresia and a tracheoesophageal fistula (EA + TEF) are a rare combined congenital anomalies associated with a high morbidity and mortality. For those patients that have survived these malformation, there has been limited long-term follow up. This case report describes a 23-year followup with evaluation of the patient's pulmonary, cardiac and gastrointestinal function.

Unilateral pulmonary agenesis is a rare congenital anomaly with a frequency estimated at 1:15,000 autopsies [1]. EA + TEF has an occurrence rate of 2500–4000 live births. The successful repair for the combination of these 2 anomalies was first reported in 1985 [2] with a 5 year followup with no respiratory symptoms. Additional reports of long-term survivors have followups limited to 12–28 months [3–5]. This case report extends the followup period to early adulthood to follow potential changes as growth occurs.

1. Case report

The patient was a 4.45 kg term female delivered by Caesarian section for a transverse position with Apgars 5 and 7 at one and 5 min of life respectively. She initially required supplemental oxygen by nasal cannula. A one-view chest x-ray demonstrated an opacified right hemithorax (Fig. 1). She was transferred to our neonatal unit for further management. She developed a decreasing PO_2 on 100% O_2 immediately after arrival. An echocardiogram (ECHO) demonstrated a VSD and ASD defects with normal myocardial function. A nasal catheter was inserted and did not pass beyond T2 with air in the GI tract consistent with a distal TEF. No other anomalies were noted on the admitting physical exam. The patient was intubated and managed with hyperventilation to correct her persistent fetal circulation, She developed multiple episodes of bradycardia and desaturation associated with an obstructed endotracheal tube. An emergency rigid bronchoscopy was performed in the neonatal unit demonstrating a large, high TEF and an absent right bronchus. The concern was that the large fistula was creating significant reflux responsible for the deteriorating pulmonary status. A #3 French Fogarty catheter was placed in the fistula and occluded. The patient was then taken to the operating room for an immediate repair. She underwent a right thoracotomy with a primary end-to-end anastomosis with mild tension. A Stamm gastrostomy was also performed. A barium swallow on postoperative day 7 revealed no anastomotic leak and mild reflux. The patient was discharged home at 4 weeks with no oxygen requirement. She received supplemental gastrostomy feedings, which were continued until 8 months of age secondary to her gastrointestinal reflux. Except for a hospitalization at 4 months of age for RSV, there were no further interval imaging or diagnostic studies.

2. Result

The patient is now 23 years old with no evidence of cardiopulmonary limitation. She continues to have reflux like symptoms but takes no prescribed medications except for tums as needed. She is presently enrolled in an exercise program with no significant limitations. A recent medical evaluation included a chest x-ray with opacification of the right hemithorax with marked hyperinflation of

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Fig. 1. Newborn chest x-ray.

the left lung with a mediastinal shift to the right. Minimal scoliosis is present (Fig. 2). An ECHO revealed an ejection fraction >55% and no evidence for pulmonary hypertension. In addition, no septal wall defect was present. Pulmonary function studies were consistent with mild obstruction with a FEV1/FVC at 69% predicted. After correcting for the alveolar volumes, the patient has no evidence for a diffusion defect.

3. Conclusion

Unilateral pulmonary agenesis is compatible with long term survival if no significant cardiovascular, gastrointestinal or genito-urinary malformations are present [6]. Previous reports describe distortion of the trachea and aortic arch due to severe mediastinal shift contributing to a poor prognosis [7]. An expandable implant in the chest has also been described in this group of patients [1].

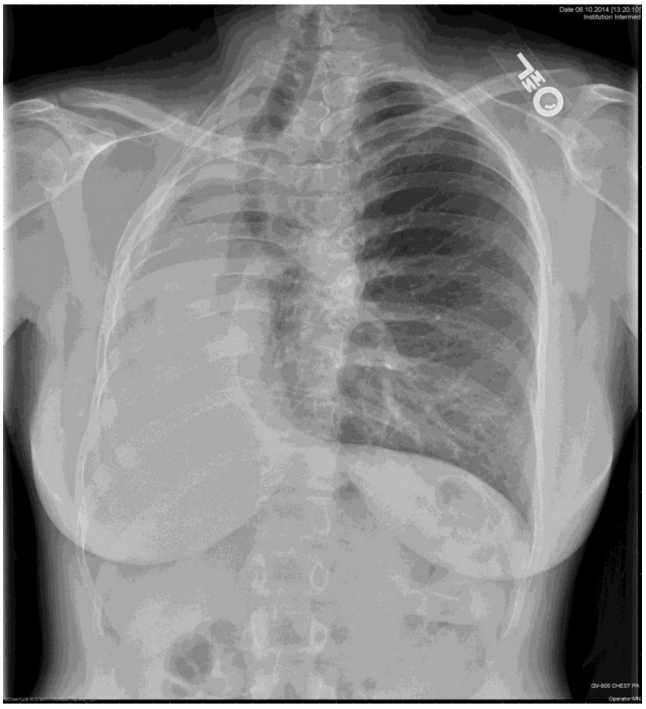

Fig. 2. Chest x-ray at 23 years.

Implants have been used in older children and adults after a pneumonectomy but have less applicability in the newborns who have compensated for their shifted anatomy. Successful management is achieved with this combination of anomalies if medical and surgical care is carefully planned and associated anomalies corrected.

Conflicts of interest

The authors declare they have no conflicts of interest.

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